Diabetic Papillopathy: Clinical Features & Fluorescein Angiographic Findings

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Abstract: Aim of the work: Description of clinical course and fluorescein angiographic features of diabetic papillopathy**. Methods:** Prospective case series. **Results:** Nine eyes of six patients met the study definition of diabetic papillopathy. Patients were in the sixth decade (mean age, 56.5 years). All patients had type II Diabetes. Patients presented with hyperemic disc swelling, and symptoms and signs of optic nerve dysfunction. Fluorescein angiography demonstrated leakage from telangiectatic optic nerve head (ONH) vessels but with no capillary non-perfusion (CNP) in any case. Disc swelling & leakage on fluorescein angiography resolved on average within 2.9 months. BCVA improved in five eyes (55%), remained stable in three (33%), and deteriorated in one eye (11%). Optic disc neovascularization (NVD), and consecutive optic atrophy were the encountered complications. **Conclusion:** Diabetic papillopathy is a relatively benign syndrome of transient optic disc swelling that occurs in diabetic patients. It is distinct from NA-AION. This entity should be always considered a diagnosis of exclusion in diabetic patients with disc swelling, variable visual acuity, normal blood pressure, with no signs of increased intra-cranial pressure.

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1- Introduction

A distinct syndrome of transient optic disc swelling associated with minimal deterioration of visual function, especially in young population with type I diabetes mellitus, was first described by Lubow and Makley in 1971. They considered it a milder variant of non-arteritic anterior ischemic optic neuropathy 'NA-AION'[1]. Later studies concluded that this syndrome is a distinct entity mimicking NA-AION though clinically distinguishable from it by the paucity of symptoms and signs of optic nerve dysfunction, and remission without significant sequelae. The term 'Diabetic Papillopathy' was introduced[2,3].

The current consensus of the clinical profile of diabetic papillopathy patient is a long standing diabetes mellitus regardless of patient's age, varying type and severity of diabetic retinopathy, mild to moderate vision loss, minimal symptoms & signs of optic nerve dysfunction.

Ophthalmoscopy reveals unilateral/ bilateral hyperemic disc swelling, peri-papillary intra-retinal hemorrhage, radially – oriented telangiectatic vessels extending on the disc surface & not extending into the vitreous cavity.

Fundus fluorescein angiography demonstrates absent extensive CNP areas in the central fundus. Early phases will clearly demonstrate the dilated radially-oriented ONH capillaries that later leak fluorescein into the edematous tissue of the disc and peri-papillary retina without leakage into the vitreous cavity or obscuration of retinal vessels. In later frames retinal vessels no longer containing dye are shadowed against the intense hyperfluorescent background at a deeper plane giving rise to characteristic silhouetting sign. Visual field will be normal, minimally affected or shows enlarged blind spot. The condition usually runs a benign course that lasts less than one year, with spontaneous recovery of disc edema albeit mild palor, and reversal of visual field changes. Visual prognosis is good[4-8].

No specific therapy is indicated beyond adequate control of diabetes mellitus, though some authors have tried corticosteroids (systemic, peri-ocular), heparin low molecular weight dextran but with no clear cut efficacy[1,2,9].

Theories of pathogenesis of diabetic papillopathy implicate diabetic micro-angiopathy. Some investigators suggest that the disc swelling may represent a superficial retinal vascular disturbance with transient leakage of fluid into and around the optic nerve head. Others believe that the disc swelling is related to deeper optic nerve head vascular compromise and axoplasmic flow disruption[1,10].

Diabetic papillopathy should be always considered a diagnosis of exclusion in diabetic patients, with disc swelling, variable visual acuity, minimal signs of optic nerve dysfunction, normal blood pressure, with no signs of increased intra-cranial pressure[5,7,11].

2. Patients and Methods.

A prospective case series conducted in the Retina Department of the Research Institute of Ophthalmology, Egypt during the year 2010.

Inclusion criteria included all diabetic patients of any age group with unilateral/bilateral disc swelling less than 12 months duration regardless of the type of diabetes mellitus.

Exclusion criteria were patients with deranged renal parameters, severe systemic hypertension (Systolic ≥ 180, diastolic ≥ 110), abnormal neuro-imaging, unexplained erythrocyte sedimentation rate> 40 mm/h to exclude Giant Cell Arteritis (GCA), NVD, optic nerve head palor, significant dyschromatopsia, major field abnormalities, afferent pupillary defect, non-improving visual acuity in absence of macular disease, and disc edema due to other pathologies (vasculitis, infectious, inflammatory).

Examination of all patients included full history, BCVA (Snellen notation), slit lamp examination including fundus biomicroscopy, applanation tonometry, and indirect ophthalmoscopy. All patients had visual field examination using Humphrey automated perimetry & fluorescein fundus angiography. Diabetic retinopathy grading was done according to the criteria of ETDRS. Average follow-up period was 2.9 months.

3. Results

Nine eyes of six patients were studied, Table 1.Five females and one male. All patients were in the sixth decade of life; average age 56.5 years. All patients had type II diabetes mellitus and all had deranged HbA_1C. Presenting symptoms included painless blurred vision, transient visual obscuration,& positive scotoma or all of them. Average duration of symptoms was 1.4 months. Initial BCVA ranged from 20/400-20/32 with 1/3 of patients scoring ≤ 20/200. Ophthalmoscopy revealed hyperemic disc swelling, peri-papillary splinter hemorrhage, and radially-oriented telangiectatic vessels extending over the disc surface, Figure 1.Fluorescein angiography in all cases revealed non-proliferative diabetic retinopathy (NPDR), absent extensive capillary non-perfusion areas in the posterior pole, leakage of dye into the edematous tissue of the disc and the peri-papillary retina (with no leakage into the vitreous cavity and without obscuration of the major retinal vessels). Characteristic silhouetting was evident in late frames, Figure 2. Visual field findings included arcuate scotoma and enlarged blind spot.

At the end of follow-up period BCVA improved in five eyes (55%), remained stable in three eyes (33%), and deteriorated in one eye (11%), Figure 3.

Seven eyes had spontaneous resolution of disc edema, with reversal of fundus and fluorescein angiographic findings. One eye, which had non-proliferative diabetic retinopathy at the time of recruitment, developed florid NVDs. Another eye developed consecutive optic atrophy, Figure 4.

Table 1. Patients' Characteristics

Patient Characteristics												
N	Sx	A	Eye	DM			Optic Disc	DR	BCVA		Follow-up	Complications
				T	D'yrs.'	HbA1c	Edema	Grade	Initial	Final	Months	
1	F	60	OS	II	5	9.10%	Diffuse	NPDR	20/40	20/30	4.5	None
2	F	58	OD	II	10	8.20%	Diffuse	NPDR	20/50	20/50	1	None
3			OS				Diffuse	NPDR	20/400	20/100		None
4	F	52	OD	II	8	11.10%	Diffuse	NPDR	20/400	20/400	6	COA
5	F	61	OD	II	10	7.80%	Diffuse	NPDR	20/80	20/30	2	None
6			OS				Diffuse	NPDR	20/200	20/80		None
7	M	53	OD	II	0.25	6.50%	Diffuse	NPDR	20/100	20/80	2	NVDs
8			OS				Diffuse	NPDR	20/50	20/80		None
9	F	55	OD	II	12	8.50%	Diffuse	NPDR	20/100	20/100	2	None

N=Name; Sx=Sex; A=Age; T=Type; D=Duration; DM=Diabetes Mellitus; DR=Diabetic Retinopathy; COA=consecutive optic atrophy; NVDs=Neovascularization of the disc; NPDR=Non-proliferative diabetic retinopathy

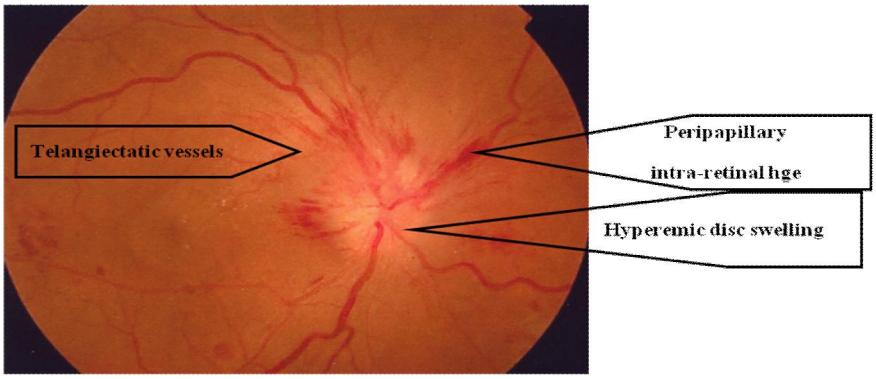
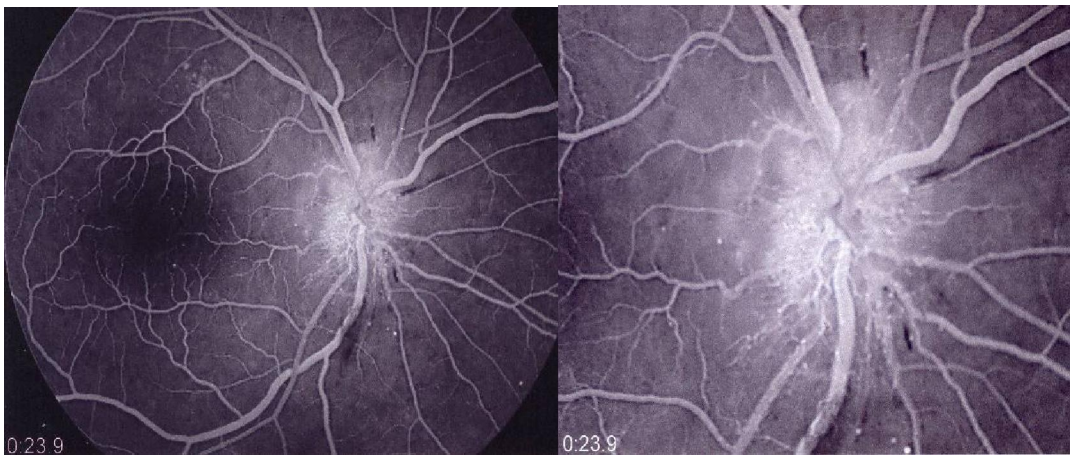
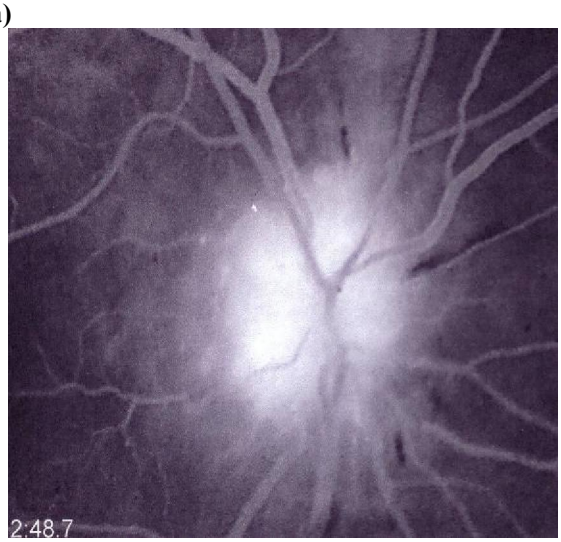


Figure 1. Fundus appearance of the ONH of a patient with diabetic papillopathy. Note that the telangiectatic vessels are radially oriented, and not extending into the vitreous cavity unlike NVDs.



(a) **(b)**



(c)

Figure 2a. Venous phase fluorescein angiography in a diabetic papillopathy case shows complete absence of CNP areas in the posterior fundus. **Figure 2b**. Close-up of the ONH seen in figure 2a, Fluorescein angiography clearly demonstrates the radial orientation of the telangiectatic vessels that leak dye into the disc tissue & the peri-papillary retina but without leakage into the vitreous cavity or obscuration of the major retinal vessels. **Figure 2c**. Later frames of the same patient demonstrating retinal vessels no longer contain dye and are shadowed against the hyperfluorescent background at a deeper plane giving rise to characteristic 'silhouetting appearance'.

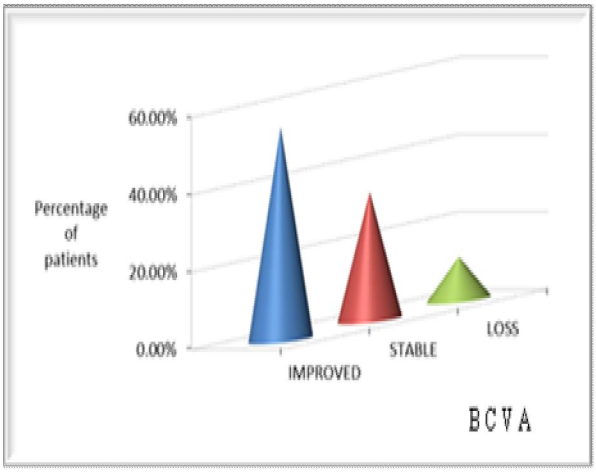


Figure 3. BCVA at end of follow-up period.

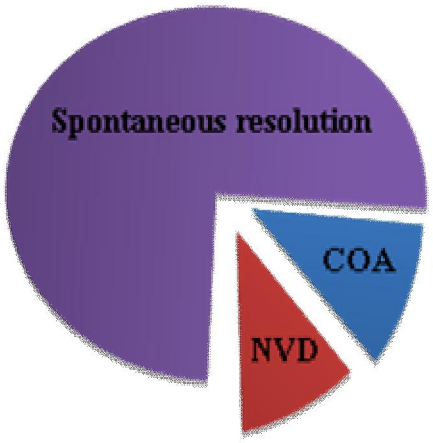


Figure 4. Complications at end of follow-up period. (NVD = Neovascularization of the Disc), COA = Consecutive Optic Atrophy

4. Discussion

The sample size in our work is too small to obtain statistical significance, though we can compare our findings with those published in literature by other authors.

In our series ophthalmoscopy revealed hyperemic disc swelling, peri-papillary intra-retinal hemorrhage along with radially-oriented telangiectatic vessels extending over the disc surface without extension into the vitreous cavity as in the case of NVDs. Fluorescein angiography revealed absent capillary non-perfusion areas in the posterior pole, leakage of dye into the disc tissue and the peri-papillary retina (unlike NVDs which leak the dye profusely into the vitreous cavity and cause obscuration of the major retinal vessels).Automated perimetry showed arcuate scotoma or enlarged blind spot. These findings are in accordance with clinical signs, fluorescein angiographic findings and visual field changes reported by other authors[2-7].

In contrast to published reports which stated that diabetic papillopathy patients usually present with mild to moderate visual loss; we had three eyes with severe visual loss (scoring ≤ 20/200 on Snellen chart) at presentation. This late finding was also reported by Regillo et al. who stated that diabetic papillopathy patients could present with profound visual loss[4].

In terms of complications, the majority of eyes in our series had spontaneous resolution and improvement of visual acuity. This comes in accordance with published literature which states that the condition usually runs a benign course[2,3]. In contrast to this notion one of our patients developed irreversible visual loss due to consecutive optic atrophy. Another patient who had NPDR at the time of enrollment developed florid NVDs &needed pan retinal photocoagulation. This late finding has been mentioned by other authors who reported diabetic papillopathy patients progressing to PDR[12,13].

Conclusion

Diabetic papillopathy is a real entity, yet to be well defined through larger scale prospective randomized controlled trials. It is noteworthy that this entity should be always considered a diagnosis of exclusion in diabetic patients, with disc swelling, variable visual acuity, minimal signs of optic nerve dysfunction, normal blood pressure, with no signs of increased intra-cranial pressure.

Ophthalmologists should be able to recognize the problem early so as to spare the patient un-necessarily, invasive and extensive neurological evaluation, and to closely observe those patients for the development of NVDs.

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